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Evaluation of Neurological and Auditory Development in Children with Congenital Heart Disease using Essence Q Questionnaire and Auditory Brainstem Response (ABR) Test

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Asadollah TANASAN MD [1],
Fatemeh EGHALIAN MD [1],
Helen BEHMANESH MD [2],
Salman KHAZAEI MD [3],
Farhad FARAHANI MD [4],
Firozeh HOSSEINI MD [1]

1-Department of Pediatrices, Hamadan University of Medical Sciences, Hamedan, Iran
2- Departmentof Psycology, Hamadan University of Medical Sciences, Hamedan, Iran
3-Research Center for Health Sciences,Hamadan University of Medical Sciences, Hamedan, Iran
4-Depatment of Ear, Noise and Throath, Hamadan University of Medical Sciences, Hamadan, Iran

Corresponding Author
Hosseini F. MD
Email: firozeh_hosseini@yahoo.com
Department of Pediatrices, Hamadan University of Medical Sciences, Hamedan, Iran

Abstract

Objectives

The progress of cardiac surgery in children and the increase in the survival of children with Congenital Heart Disease (CHD) has led to consider another issue called a neurodevelopmental disorder. In this study, 53 children with CHD were evaluated in terms of development with the Essence Q questionnaire, Otoacoustic Emission (OAE), and Auditory Brainstem Response (ABR) regarding these patients' hearing and risk factors. The Essence Q scores were also examined.

Materials & Methods

In this prospective, cross-sectional study, the researchers included 53 children diagnosed with CHD. Initially, each child underwent ABR and OAE tests. Subsequently, data on potential risk factors associated with neurodevelopmental delay were collected. A trained project associate administered the Essence Q questionnaire, using parents' information as a guide. Following data collection, this study proceeded with an in-depth analysis of the information.

Results

Thirty-six boys (67.92%) and 17 girls (32.08%) with CHD were included in the study. The mean age of children was 2/23+_8.11 months. The mean Essence Q score for boys was 7.48+_2.57. Moreover, the average score for girls was 2.23 8.11. According to this questionnaire, 39 patients (73.58%) had hyperactivity disorder, 46 patients (86.79%) had behavioral disorders, and ten patients (16.98%) had a motor delay. Unlike previous studies, all patients had normal OAE and ABR hearing.

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Conclusion

This study demonstrated that factors such as developmental delay in the first year, a known genetic disease, and a history of seizures significantly impacted the Essence Q score. However, elements like prematurity, the use of ventilation, abnormalities on the dorsum, and the number of days post-surgery did not significantly affect the Essence Q score. Essence Q can be a reliable tool in screening for neurodevelopment in children with CHD.

Keywords: Congenital heart disease, Neurodevelopmental Delay, ESSENCE Q questioner, Auditory, Brainstem Evokes Potential
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Introduction

Due to significant advances in heart surgery and pre/post-operative care, there has been a noticeable decrease in the mortality rate of children with heart disease. This includes children suffering from complex heart diseases, who now have an increased chance of survival. As a result of this reduction in mortality, physicians and researchers have shifted their focus toward evaluating long-term complications in these children, aiming to develop effective treatment strategies to prevent further complications (1). Among the most common complications in these children, a well-proven complication is a developmental delay. Previous reports have shown that up to 50% of these children can have developmental delays (2). The pattern of developmental disorders in these children is described as mild cognitive information-social communication deficits and communication skills, including language and attention and impulsive behaviors and actions in children with Congenital Heart Disease (CHD). Many theories have been proposed about the cause of developmental delay in these children. Biological factors such as dysgenesis expressed in the brain and heart (3,4), changes in cerebral blood flow in the uterus, brain structure abnormalities, and brain immaturity are expressed before heart surgery (5). Other factors, such as physiological defects of the heart and surgical and pharmacological treatments or underlying genetic diseases, should also be considered (6,7).

In addition to the listed factors, environmental factors and interventions can reduce and increase this risk (8). In addition, children with CDH are also prone to hearing impairment, delaying the acquisition of speech skills and speech delay (9). Of these studies, one old study estimated the prevalence to be 16% in patients with CHD (10). Furthermore, a recent study estimated the prevalence to be as high as 28.6% among childhood survivors after the Norwood/Sano operation only (11). Besides, another study showed SNHL prevalence in asymptomatic, palliated/repaired patients with CHD followed in our high-risk clinics and undergoing routine surveillance was

11.6% (9).

To achieve this goal, we must detect developmental delays in patients early because early detection allows us to intervene quickly and start advanced rehabilitation services such as physiotherapy, speech therapy, and occupational therapy for the patients quickly and help improve development. Different tools are used to assess and screen children's development on an outpatient basis. Parents or children mostly fill out these questionnaires. These questionnaires screen for difficulties, such as the need for highly trained and specialized people that cannot be done in an outpatient center. Among the new screening tools since 2010, the Essence Q questionnaire was introduced by Gilbert et al. (11). The ease of working with this questionnaire has led to several research studies on children using this questionnaire. In this study, the factors of developmental delay and the score of the Essence Q questionnaire were examined and statistically analyzed.

Materials & Methods

This cross-sectional was carried out for a year, from May 2020 to May 2021. The research site was the Development Clinic Center of Besat Hospital, a third-class tertiary referral hospital in Hamadan city in western Iran.

Inclusion criteria:

Following parental consent, each child with congenital heart disease (CHD) was referred to a cardiologist. Initially, all participants underwent an ABR test. Subsequent to this, information was collected and recorded in a questionnaire. The data included the child's name, age, history of prematurity and genetic disease, duration of hospitalization post-surgery, the nature of their heart disease (either congenital and cyanotic or non-cyanotic), and the patient's treatment strategy (whether surgical or drug-based).

In addition, the questionnaire captured the patient's history of using the pump during surgery, their head circumference, any history of seizures, and developmental delays experienced within the first year. Finally, the OAE and ABR test results for all patients were also documented. Then, the Essence Q questions were asked by the trained colleague of the project with the help of parents' information, and then the information was analyzed.

The Essence Q questionnaire introduced by Gilbert et al. had 12 questions about different neurodevelopmental domains. This questionnaire has shown 94% sensitivity and 53% specificity in evaluating previous studies.

Descriptive statistics, including frequency and percentage, were used to describe categorical variables and mean and SD for continuous variables. The student's t-test assessed the relation between developmental parameters and Essence Q score. Data were analyzed using SPSS software version 23. A significant level was considered less than 0.05.

Results

The present study enrolled 53 patients with CHD during the mentioned time. The number of 36 (67.92%) were boys, and the rest were girls. The mean age of patients was 26.98+_10.64 months (range: 18-48 months). According to the Essence Q questionnaire, the results of 12 questions are shown in Table (1).

Essence Q score was 7.47+_2.57 for boys and

Table 1: Response status to the 12 questions of the Essence Q questionnaire

Question	Yes	Somewhat	No
Q1.General developmental.	35 (66.04)	9 (16.98)	9 (16.98)
Q2.Motor developmental milestones	31 (58.49)	13 (24.53)	9 (16.98)
Q3.Sensory reactions (touch, sound,…)	52 (98.11)	0	1 (1.89)
Q4.Communication/Language/babble	46 (86.79)	5 (9.43)	2 (3.77)
Q5.Activity/Overactivity/passivity/impulsivity	7 (13.21)	7 (13.21)	39 (73.58)
Q6.Attention/Concentration/Listening	50 (94.34)	2 (3.77)	1 (1.89)
Q7.Social interaction/interesting	45 (84.91)	7 (13.21)	1 (1.89)
Q8.Behavior	51 (96.23)	1 (1.89)	1 (1.89)
Q9.Mood	1 (1.89)	6 (11.32)	46 (86.79)
Q10.Sleep	45 (84.91)	6 (11.32)	2 (3.77)
Q11.feeding	33 (62.26)	14 (26.42)	6 (11.32)
Q12.Funny Spells/absence	0	3 (5.66)	50 (94.34)

Table 2: Relation between developmental parameters and Essence Q score

Variable	Yes	No	Value
Premature	8±2.31	7.65±2.5	0.79
Motor delay	8.29±3.11	7.17±1.67	0.1
Genetic problem	10.75±2.87	7.43±1.93	0.008
Underwent ventilator after surgery	7.35±1.61	8.26±3.51	0.20
Having seizures	8.44±4.22	7.52±1.98	0.31
Normal head circumference	7.67±2.49	8.0±2.83	0.85

8.11+_2.23 for girls (P=0.18). The average score of Essence Q was significantly higher in children aged 18 to 24 months than in children 36-48 months (8.29+_2.71 vs. 7.05+_1.87, P=0.008).

The relation between developmental parameters and Essence Q score is presented in Table 2. The number of four patients was premature. No significant relationship was observed between prematurity and delay in working with Essence Q score (P>0.05). A significant relationship existed between known genetic diseases such as Down syndrome and Essence Q score (P=0.004). Thirty-four patients underwent a ventilator after surgery, and 19 did not need a ventilator. No significant difference between ventilator use and Essence Q score (P=0.2) was found. The mean score of Essence Q in patients with surgery was 7.35+_1.61, and in non-surgery patients, it was 8.26+_3.51, which

this difference was not statistically significant (P= 0.02). Forty-five patients had normal head circumference, and 38 patients had abnormal head circumference, which was not significantly different in Essence Q score (P = 0.59). The mean number of hospitalization days after surgery was 13.13+_14.07 (0 days to 60 days), and there was a negative correlation between the number of hospitalization days after surgery and Essence Q score, but it was not significant (r=-0.16, P=0.26). Nine patients had seizures after heart surgery, and 44 did not. Accordingly, the difference in Essence Q score between the two groups was significant (P=0.31).

Twenty-two patients had CHD, and 31 patients had simple cyanotic heart disease. Forty-one patients had a history of using a pump in surgery, and 12 patients did not have a history of using a pump in surgery. Fifty-three patients had normal OAE and ABR.

Discussion

This study showed that the Essence Q questionnaire is an effective tool for screening neurodevelopmental delays in children with CHD. We discovered that genetic disorders and seizures significantly impact the Essence Q scores. In contrast, factors such as prematurity, post-surgery ventilator usage, motor delays, and head circumference did not significantly influence the Essence Q scores. In recent years, increasing the survival of children with CHD has been made possible through new cardiac surgery techniques and pre-and post-surgery care. Decreased mortality has led to the survival of these children, and the main complication in these children, which is Developmental Delay, is receiving more attention. The cause of developmental delay in these children is multifactorial, and in addition to the numerous causes that have been raised, even the genetic background has been considered (12).

Now, waiting and watching have been replaced by quickly finding developmental delays in these children and taking quick action. As a new tool, the Essence Q questionnaire has drawn attention recently among different ways of assessing developmental delay. This questionnaire has shown 94% sensitivity and 53% specificity in the evaluation of previous studies (13), so with high sensitivity, it can be a good tool in screening patients for developmental delay.

Previous studies have identified many factors such as prematurity, genetic syndromes, history of the mechanical ventilator for breathing or resuscitation and seizures after surgery, history of microcephaly and brain dysgenesis defects, and length of hospital stay of more than two weeks as interfering factors in the development of these children (14).

This study showed that in children with CHD, developmental delay in the first year, genetic problems, and history of seizures after surgery significantly affected the Essence Q score. However, the child's prematurity, a history of ventilator use, abnormal head circumference, and several hospital stays after surgery had no significant effect on the Essence Q score.

The remarkable point of this study was that 73.58% of the patients had warning signs of hyperactivity, and 86.79% of the children had mood problems based on this questionnaire. A recent study of behavioral disorders, especially depression and anxiety in children with CHD under ten years of

age, found that 16% of children aged 4 to 9 years had symptoms of depression and anxiety (15). Notably, the age of children in this study was less than in the above mentioned study. Correspondingly, 29% of motor hyperactivity symptoms were reported in children aged seven to 15 years, but at the age of five to ten years, 30% of these children had symptoms of hyperactivity and lack of attention and concentration (16). In the present study, the symptoms of hyperactivity were reported by parents at a much younger age than in this study. However, the symptoms of hyperactivity may be somewhat exaggerated by the parents. Movement delay is also noticeable in these children. Seemingly, the parents were not justified in stereotyped, and many regular movements of the child were classified as stereotyped movements. Otherwise, hearing loss has a negative effect on speech delay. Several hypotheses support inner ear ischemia and hypo-perfusion after open heart surgery as the cause of hearing loss in patients with CHD (16, 17). Ischemia or hypo-perfusion can be a result of hypoxia during surgery, bolus administration of furosemide after cardiopulmonary bypass surgery (11), long duration of ECMO therapy and clinical seizure activity prior to ECMO (18). This study evaluated patients for possible auditory impairments by ABR and OAE tests, although none developed hearing impairment. Likewise, the current research aimed to evaluate the effect of hearing impairment on speech delay, although it was unsuccessful since none of the cases had impaired hearing tests. Previous studies have reported the rate of hearing impairment to be from 11.6% to 28.6%, but surprisingly, all of our patients had normal ABR and OAE tests. Failing to find abnormal ABR results in this study suggests that individuals with complex cardiac defects may not survive long enough to be included in this analysis. However, in hospital settings equipped with experienced pediatric cardiac surgery teams and advanced support facilities, patients with such defects may have improved survival rates. Notably, these patients may still develop hearing impairment as they age. Further studies with a larger sample size in referral pediatric cardiology centers are recommended to evaluate the precise rate of hearing impairments in patients with congenital heart defects.

In Conclusion

The Essence Q questionnaire can be used as a suitable screening tool for children with CHD. Hyperactivity and mood disorders with this questionnaire are significant in these children.

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This study was approved by the Ethics Committee of Hamadan University of Medical Sciences, and information was obtained verbally from parents

Author's contribution

Dr Esfehani conceived and designed the analysis. Dr Sahraii collected the data. Dr Seif Rabie contributed data or analysis tools and performed the analysis. Dr hosseini wrote the paper. Dr Sedigi

and Dr Esfehani performed revision of the paper.

Conflict of interest

The authors declare that they have no conflict of interest. IR.UMSHA.REC.1396.831

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